Radiology Corner

Superior Vena Cava Syndrome: Radiological Diagnosis and Endovascular Treatment

Guarantor: CPT P. Gabriel Peterson, M.D., USA[1]
Contributors: CPT P. Gabriel Peterson, M.D., USA[1]; Joseph Ronsivalle, D.O.[2]; COL Les Folio, USAF, MC, SFS [1,3]

Note: This is the full text version of the radiology corner question published in the February 2008 issue, with the abbreviated answer in the March 2008 issue.

SVC syndrome results from increased hydrostatic pressure in the venous system distal to the SVC, through which blood flow has been diminished or obstructed. The source for obstruction may be an extrinsic process compressing the SVC or an intraluminal process, such as a venous thrombosis. The diagnosis is most often confirmed with computed tomography or venography. Diagnostic criteria include the demonstration of obstruction as well as the opacification of collateral venous systems. The following case demonstrates the most common radiographic findings of SVC syndrome as well as several therapeutic options available. Finally, important radiographic landmarks consistent with pathology are demonstrated throughout the case.

Introduction

William Hunter, a Scottish physician who gained notoriety with regard to his description of aneurysms (1), described the first case of SVC syndrome in 1757, which was caused by a syphilitic aortic aneurysm (2). It was originally thought that the majority of cases were caused by aneurysms, however, extrinsic compression from malignancy is now the most common etiology, with bronchogenic lung cancer and lymphoma representing 80-97% of cases (3).

History

A 43 year old female presented with a five-day history of bilateral facial, neck, and upper extremity swelling. She had a history significant for poorly differentiated infiltrating ductal carcinoma, which was diagnosed and subsequently treated with a right modified radical mastectomy approximately four months prior. A right-sided indwelling catheter had been in place for approximately three to four months and the patient was status-post 6 cycles of doxorubicin and cyclophosphamide (AC) chemotherapy.

Summary of Image Findings

Conventional radiography typically reveals a widened superior mediastinum. In this case there is also a bulge in the region of the azygos vein (Fig 1).

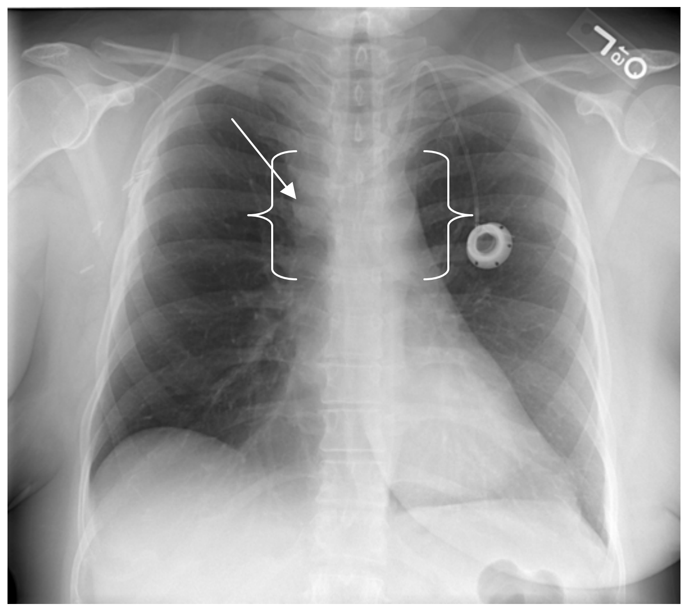

Fig. 1 Frontal radiograph of the chest, which demonstrates absence of the right breast shadow, left-sided mediport with catheter tip extending to the region of the SVC, and widened superior mediastinum (bracket) with bulge in the region of the azygos vein (arrow).

Computed tomography images revealed a filling defect consistent with venous thrombosis in the superior SVC at the level of the left brachiocephalic vein. The catheter tip is visible in the filling defect (Fig 2). Initial contrast venography revealed complete obstruction (Fig. 3).

Patient Discussion

After an initial infusion of tissue plasminogen activator (tPA) an infusion catheter was passed across the length of the thrombosis and tPA was infused overnight. The following day revealed additional contrast material bypassing the thrombosis into the SVC (Fig. 4).

The catheter was repositioned and irrigation was continued. On day 3 the infusion catheter was removed and a wire was passed over the thrombus for balloon angioplasty. Lastly,

[1] Diagnostic Radiology; Walter Reed Army Medical Center, Washington, DC 20307
[2] Department of Radiology, Madigan Army Medical Center, Fort Lewis, WA
[3] Department of Radiology and Radiological Sciences; Uniformed Services University of the Health Sciences, Bethesda, Maryland 20814-4799

Report Documentation Page

Form Approved
OMB No. 0704-0188

1. REPORT DATE	2. REPORT TYPE	3. DATES COVERED
MAR 2008		**00-00-2008 to 00-00-2008**

4. TITLE AND SUBTITLE
Superior Vena Cava Syndrome: Radiological Diagnosis and Endovascular Treatment

5a. CONTRACT NUMBER
5b. GRANT NUMBER
5c. PROGRAM ELEMENT NUMBER

6. AUTHOR(S)

5d. PROJECT NUMBER
5e. TASK NUMBER
5f. WORK UNIT NUMBER

7. PERFORMING ORGANIZATION NAME(S) AND ADDRESS(ES)
Uniformed Services University of the Health Sciences,Department of Radiology and Radiological Sciences,4301 Jones Bridge Road,Bethesda,MD,20814

8. PERFORMING ORGANIZATION REPORT NUMBER

9. SPONSORING/MONITORING AGENCY NAME(S) AND ADDRESS(ES)

10. SPONSOR/MONITOR'S ACRONYM(S)

11. SPONSOR/MONITOR'S REPORT NUMBER(S)

12. DISTRIBUTION/AVAILABILITY STATEMENT
Approved for public release; distribution unlimited

13. SUPPLEMENTARY NOTES

14. ABSTRACT

15. SUBJECT TERMS

16. SECURITY CLASSIFICATION OF:			17. LIMITATION OF ABSTRACT	18. NUMBER OF PAGES	19a. NAME OF RESPONSIBLE PERSON
a. REPORT	b. ABSTRACT	c. THIS PAGE			
unclassified	**unclassified**	**unclassified**	**Same as Report (SAR)**	**3**	

mechanical thrombectomy was performed using an Amplatz, vortex-based device (Fig. 5).

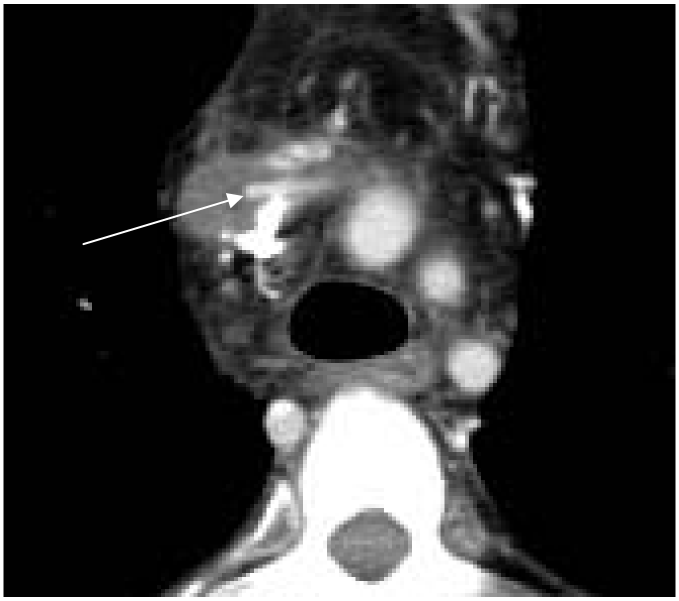

Fig. 2 Axial CT image (magnified view) after intravenous contrast administration at the level of the superior SVC with a homogeneous filling defect in the lumen of the SVC. The catheter tip is visible in the filling defect (arrow).

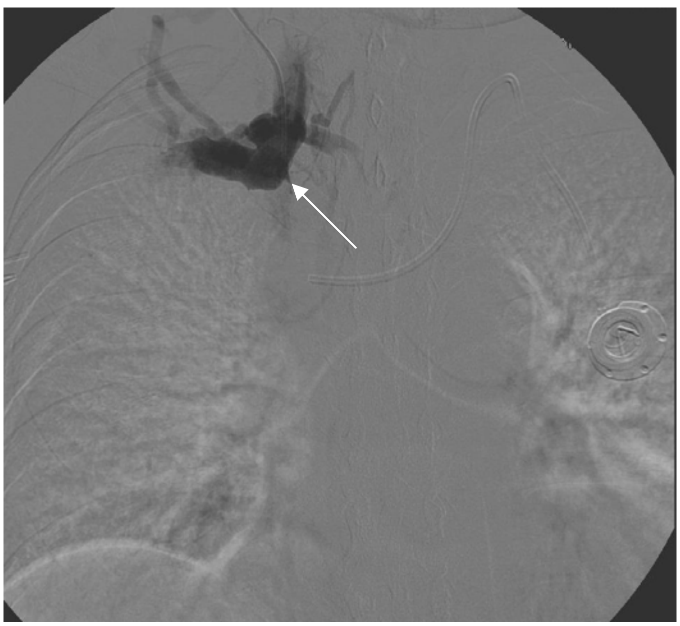

Fig. 3 Contrast venography of the SVC reveals complete obstruction. Note small strand of contrast medially outlining medial edge of thrombus (arrow), this correlates with CT finding.

Discussion

When blood flow through the superior vena cava is reduced or obstructed, backpressure naturally builds in its tributaries, namely the venous systems of the head, neck, and upper extremities. The increase in venous hydrostatic pressure causes extravasation of fluid into the interstitial tissues, which ultimately leads to the symptoms of facial and upper extremity swelling. On physical exam, there is often engorgement of superficial veins in the face, neck, and chest wall.

Fig. 4 Contrast venography demonstrates passage of contrast material distal to the thrombosis into the SVC (S). The thrombosis has irregular borders and extends from the right brachiocephalic vein (arrowhead) into the SVC across the entrance of the left brachiocephalic vein containing the mediport catheter (arrow).

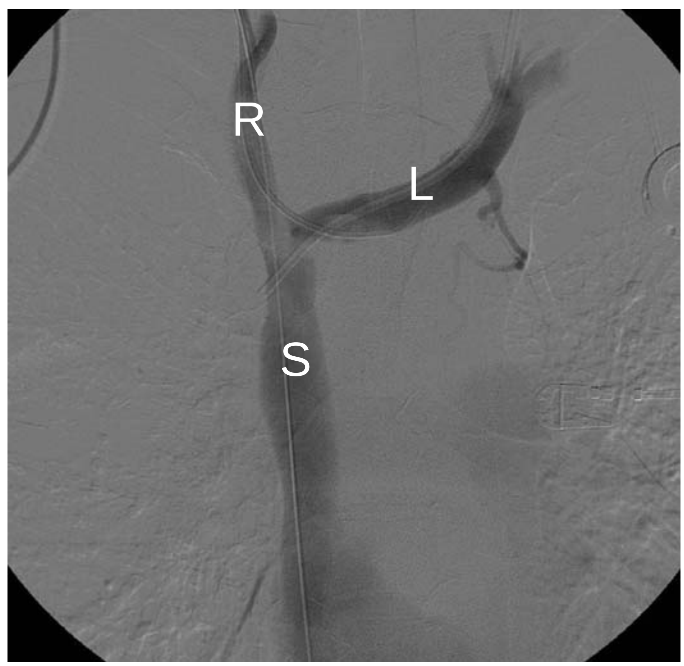

Fig. 5 Contrast venography of the SVC demonstrates complete filling of the now patent left (L) and right brachiocephalic (R) veins as well as the SVC (S).

In order for blood to return to the right atrium from the head, neck, and upper extremities when the SVC is blocked, collateral drainage routes dilate to accommodate additional blood flow. The most common collateral systems include the azygos, hemiazygos, vertebral, internal mammary, lateral thoracic, paraspinous, and esophageal venous systems.

Approximately 94% of cases are a result of bronchogenic lung cancer or lymphoma, which compress the SVC (3). As a result the initial chest radiograph is abnormal in up to 84% of cases (4). The most common findings on chest radiography include a widened superior mediastinum, evidence of a lung mass, or evidence of lymphadenopathy.

Contrast-enhanced computed tomography (CT) is an excellent modality for diagnosing the extent of obstruction. In addition, CT often allows radiologists to better characterize the underlying cause. Criteria for diagnosis include decreased or absence of opacification of central venous structures inferior to the obstruction as well as opacification of collateral venous systems (5). In one study, radiologists were able to diagnose SVC syndrome based solely on presence of collateral vessel dilation with a sensitivity of 96% and a specificity of 92% (6). Additionally, looking at the collateral venous systems as they continue into the upper abdomen can suggest the diagnosis of venous obstruction in the SVC (7).

Additional modalities for diagnosis include contrast venography, magnetic resonance (MR) angiography, and technetium-99m venography. Percutaneous contrast venography (Fig. 3-5) allows the interventional radiologist to characterize the extent of the obstruction as well as therapeutically intervene. MR angiography is appropriate for patients with contraindications to iodinated contrast (8).

Regardless of extrinsic compression or intraluminal thrombosis, balloon angioplasty with or without stent placement can often relieve the patient's symptoms within one week (9, 10). In the case of malignant compression, however, stent placement is often a palliative measure, which allows the patient to undergo necessary radiation and/or chemotherapy (11, 12). Long-term patency rates of these stents remain unknown, however, in cases of malignant etiology the stent typically lasts for the life of the patient (13).

Conversely, when the obstruction is from a benign cause, such as catheter-related thrombosis, metallic indwelling stents are avoided. If it is established that the thrombosis has occurred within five days of presentation and barring any other systemic contraindications, catheter-directed thrombolytic therapy is an option that can rapidly reduce thrombosis burden. Tissue Plasminogen Activator (tPA) has also been shown to reduce clot burden (14, 15) and can be delivered directly to the thrombosis via a multi-side hole infusion catheter advanced over the thrombus. When chronic, well-organized thrombus remains after these techniques there are several devices designed for percutaneous mechanical thrombectomy. In this case, an Amplatz, vortex-based, mechanical thrombectomy device was successful (Fig. 5). Post thrombectomy patients remain at an increased risk of reforming thrombus and are usually maintained on long-term systemic anticoagulation (16, 17).

Note: Follow this link for Category 1 CME or CNE in the MedPix™ *case of the week digital teaching file.*

http://rad.usuhs.mil/amsus.html

References

1. Friedman SG, M.D. "A History of Vascular Surgery." 2nd Ed. Blackwell Publishing, 2005; pp 31-34.
2. Hunter W. "The History of an Aneurysm of the Aorta with some remarks on Aneurysms in General." *Med Observ Inq* 1757; 1:323-57.
3. Abner A. "Approach to the patient who presents with superior vena cava obstruction. (Multimodality Therapy of Chest Malignancies: Proceedings of a Harvard Medical School Symposium)." *Chest*. Apr. 1993; 103(4): 394S-397S.
4. Parish JM, et al. "Etiologic considerations in Superior Vena Cava syndrome." *Mayo Clinic Proceedings*. Jul. 1981; 56(7):407-13.
5. Engel IA, Auh YH, Rubenstein WA, et al. "CT Diagnosis of Mediastinal and Thoracic inlet venous obstruction." *AJR*. Sept. 1983; 141(1):521-6.
6. Kim HJ, Kim HS, Chung SH. "CT diagnosis of superior vena cava syndrome: importance of collateral vessels." *AJR*. 1993; 161:539-42.
7. Bashist B, Parisi A, Frager DH, and Suster B. "Abdominal CT findings when the superior vena cava, brachiocephalic vein, or subclavian vein is obstructed." *AJR*. 1996; 167:1457-63.
8. Finn JP, Zisk JH, Edelman RR, et al. "Central venous occlusion: MR angiography." *Radiology*. Apr. 1993; 187(1):245-51.
9. Gross CM, Kramer J, Waigland J, Uhlich F, et al. "Stent implantation in patients with superior vena cava syndrome." *AJR*. 1997; 169:429-32.
10. Qanadli SD, Hajjam ME, Mignon F, et al. "Subacute and chronic benign superior vena cava obstructions: endovascular treatment with self-expanding metallic stents." *AJR*. 1999; 173:159-64.
11. Armstrong BA, Perez CA, Simpson JR, and Hederman MA. " Role of irradiation in the management of superior vena cava syndrome." *Int J Radiat Oncol Biol Phys*. Apr. 1987; 13(4):531-9.
12. Perez-Soler R, McLaughlin P, Velasquez, et al. "Clinical features and results of management of superior vena cava syndrome secondary to lymphoma." *J Clin Oncol*. Apr. 1984; 2(4):260-6.
13. Yim CD, Sane SS, Bjarnason H. "Superior vena cava stenting." *Rad Clinics of North America*. Mar. 2000; 38(2):409-24.
14. Gray BH, Olin JW, Graor RA, et al. "Safety and efficacy of thrombolytic therapy for superior vena cava syndrome." Chest. Jan. 1991; 99(1):54-9.
15. Greenberg S, Kosinski R, Daniels J. "Treatment of superior vena cava thrombosis with recombinant tissue type plasminogen activator." Chest. May 1991; 99(5):1298-301.
16. Goldberg MF, Kim HS. "Treatment of Acute Superior Mesenteric Vein Thrombosis with Percutaneous Techniques." AJR. Nov. 2003; 181:1305-7.
17. Gu X, Sharafuddin MJ, Titus JL, et al. "Acute and delayed outcomes of mechanical thrombectomy with use of the steerable Amplatz thrombectomy device in a model of subacute inferior vena cava thrombosis." J Vasc Interv Radiol. 1997; 8(6):947-5.